JACC Vol. 3, No. 3
March 1984:868–71

Primary Right Ventricular Tumor (Fibroma) Simulating Cyanotic Heart Disease in a Newborn

JOSÉ MARÍN-GARCÍA, MD, FACC, CHARLES W. FITCH, MD, RAY E. SHENEFELT, MD

Memphis, Tennessee

The clinical, echocardiographic, cineangiocardiographic and autopsy findings in a newborn infant with right ventricular fibroma are reported. The tumor caused severe right ventricular outflow obstruction simulating cyanotic heart disease. Echocardiography allowed a definitive diagnosis of the tumor mass at 10 hours of age. The clinical implications of the case are discussed.

Primary cardiac tumors, both benign and malignant, are extremely rare at all ages and occur mainly in infants and children (1–3). The clinical recognition of ventricular tumors during life may be very difficult because they may cause hemodynamic disturbances similar to those occurring in other cardiovascular anomalies, namely, those characterized by ventricular outflow obstruction (4–7). Nevertheless, with the availability of echocardiography, the localization and characterization of intracardiac tumors may be possible before operation or postmortem examination.

We describe a neonate who presented with clinical features suggestive of cyanotic congenital heart disease; however, a gigantic right ventricular tumor was demonstrated by two-dimensional echocardiography. The diagnosis was confirmed by cineangiocardiography and, subsequently, at surgery and autopsy.

Case Report

The patient, a 1 day old boy, was the product of a normal pregnancy and delivery. Apgar scores were 8 and 8 at 1 and 5 minutes and birth weight was 2.69 kg. At admission to the well baby nursery the infant was slightly hypothermic (96.6°F, rectal) and tachypneic (76/min), but examination was otherwise unremarkable. Hypothermia resolved quickly, but tachypnea persisted and cyanosis developed overnight. Chest roentgenogram at this time showed marked cardiomegaly, and the infant was promptly transferred to the neonatal intensive care unit.

From the Departments of Pediatrics and Pathology, University of Tennessee Center for the Health Sciences and Le Bonheur Children's Medical Center, Memphis, Tennessee. Manuscript received July 11, 1983, accepted September 30, 1983.

Address for reprints: José Marín-García, MD, Section of Pediatric Cardiology, Department of Pediatrics, Le Bonheur Children's Medical Center, One Children's Plaza, Memphis, Tennessee 38103.

Admission examination revealed an alert, mildly cyanotic and tachypneic infant. Vital signs included a heart rate of 172 beats/min, respiratory rate of 106/min, temperature of 98.0°F (rectal) and Doppler blood pressures of 86 and 78 mm Hg in the right and left arms, respectively, and 90 mm Hg in the lower limbs.

Cardiac examination revealed decreased intensity of the first heart sound and a single second sound. No ejection click was present. A grade 3/6 holosystolic murmur was heard maximally at the mid and lower left sternal border. A grade 2/6 mid-diastolic murmur also was audible along both sides of the lower sternum. The liver was palpable 3 cm below the right costal margin. Peripheral pulses were palpable but diminished. Admission arterial blood gas determination revealed a partial pressure of oxygen (Po_2) of 48 torr on room air. Electrocardiogram revealed right axis deviation and left ventricular hypertrophy (Fig. 1). Chest roentgenogram showed marked cardiomegaly with decreased pulmonary vascularity.

Real-time echocardiographic examination was performed at 10 hours of age using a Varian V-3400 80° phased-array sector scanner with a 3.5 MHz transducer. The image was displayed on a video monitor and recorded in real-time on 0.5 inch (1.27 cm) video cassette tape. Still-framed pictures of the video tape images were obtained for presentation. A dense cloud of echoes, filling the entire right ventricular cavity, was seen on the four chamber apical view (Fig. 2). In addition, in this view, decreased tricuspid valve excursion with posterior displacement toward a small right atrial chamber could be seen. Bulging of the ventricular septum toward the left ventricular free wall was also demonstrated. On the basis of these findings, a diagnosis of right ventricular tumor was made.

Cineangiocardiography with contrast injection into the left atrium revealed a small atrial septal defect and a "pan-

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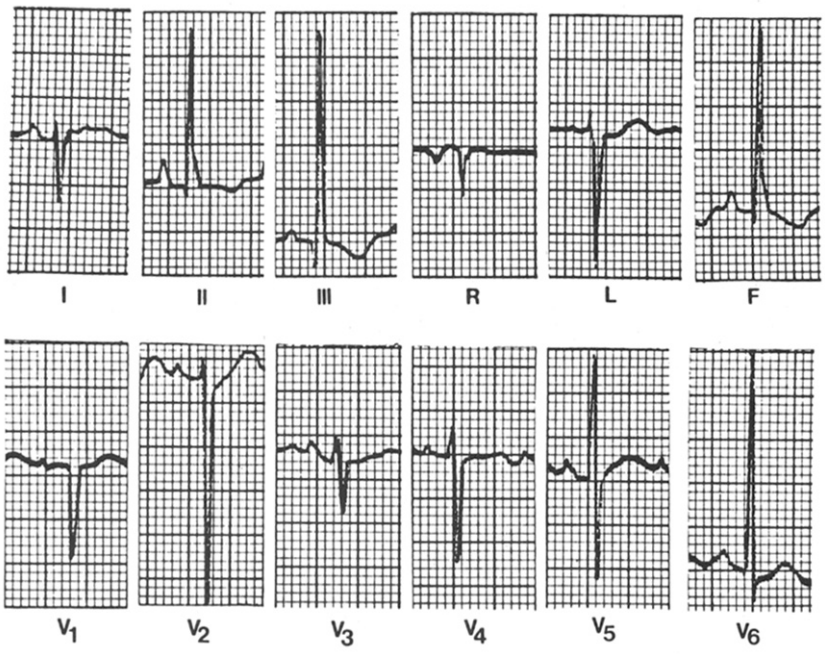
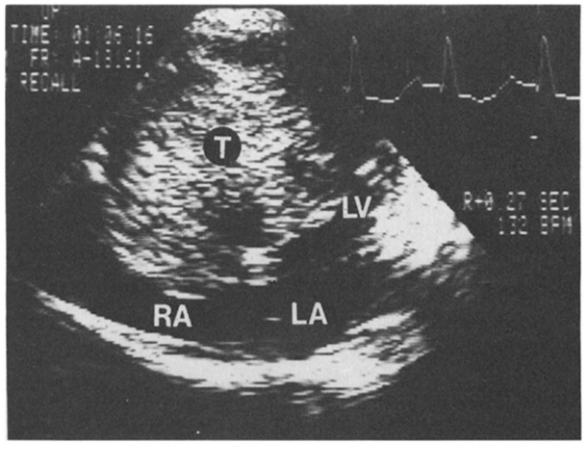


Figure 1. Electrocardiogram showing right-axis deviation and signs of left ventricular hypertrophy.

cake-like'' configuration of the left ventricular cavity (Fig. 3). A patent ductus arteriosus was also seen. Injection into the right atrium demonstrated a faint streamline flow of contrast medium to the pulmonary artery without clear delineation of the right ventricular cavity. A right to left shunt was seen across the atrial septum. After cineangiocardiography, infusion of prostaglandin E_1 was begun pending surgery.

At operation, a massive right ventricular tumor was found, occupying most of the free wall of the right ventricle and much of the interventricular septum. The extent of the tumor mass necessitated a right ventriculotomy through which much of the tumor (19.3 g) was removed, but the infant did not survive the operation.

Figure 2. Two-dimensional echocardiogram in the four chamber apical view demonstrates the tumor (T) in the right ventricle and reduced left ventricular (LV) volume. LA = left atrium; RA = right atrium.



At postmortem examination, residual tumor tissue could be seen with infiltration of the septum and right ventricular wall. The main remaining tumor masses were 3.5 × 2.4 × 1.4 and 2.2 × 2.0 × 1.0 cm. The left ventricle was displaced posteriorly, but chamber size and thickness of the free wall were within normal limits. Myocardial fibers were hypertrophied in the right ventricle but not in the left. The diagnosis of right ventricular fibroma was established by light and electron microscopic examination (Fig. 4).

Discussion

The clinical presentation in our case was compatible with cyanotic congenital heart disease, such as pulmonary valve atresia or severe pulmonary stenosis with intact ventricular septum. The systolic and diastolic murmurs were considered to be secondary to severe tricuspid valve regurgitation as may be seen in cases with pulmonary valve atresia with intact ventricular septum and normal or large right ventricle. However, in this situation, the electrocardiogram usually shows normal right ventricular forces or right ventricular hypertrophy.

Electrocardiographic findings. In our case, there were electrocardiographic findings of left ventricular hypertrophy with almost absent right ventricular forces in the right precordial leads. These findings can be interpreted in two ways. First, replacement of right ventricular free wall tissue by tumor may reduce the rightward and anterior electromotive forces generated. Forces generated by the left ventricular tissue would be subject to less cancellation by the normally powerful right ventricular forces of the young infant. Second, the presence of a relatively nonconductive intracavitary

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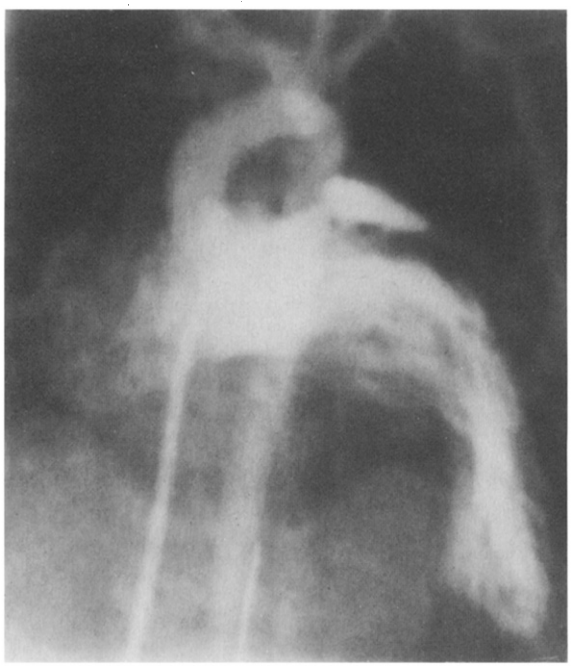
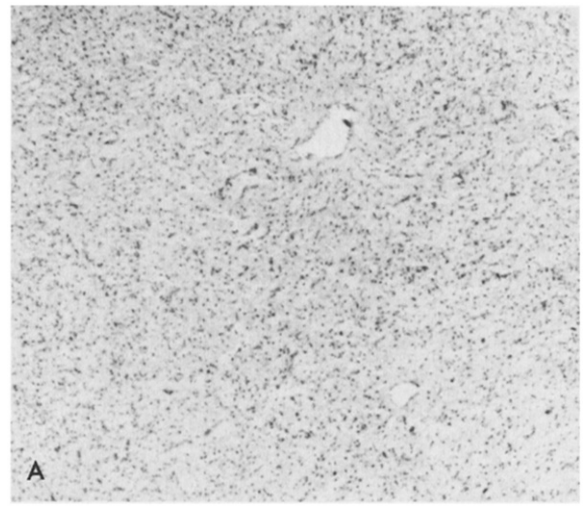


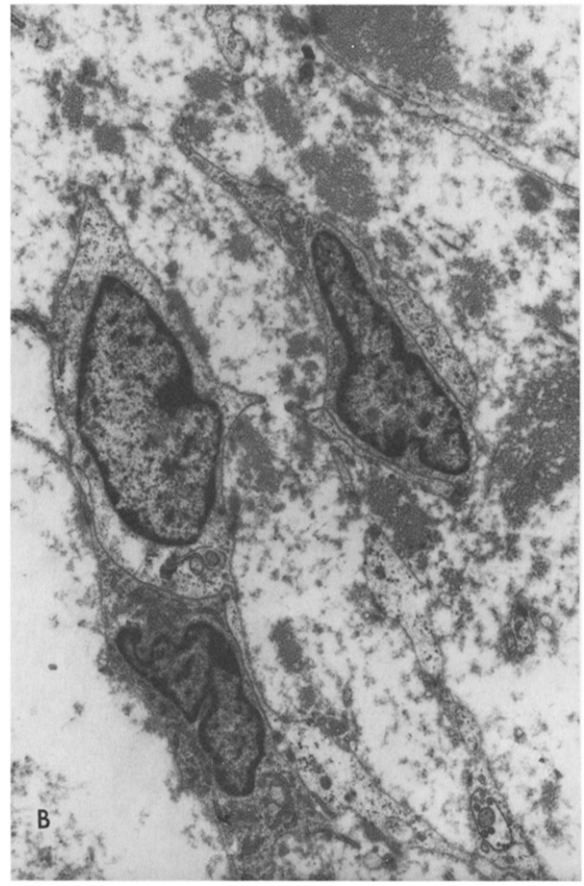
Figure 3. Left atrial angiogram showing abnormal left ventricular silhouette secondary to displacement of the ventricular septum into the left ventricular cavity by the tumor in the right ventricle.

mass, that is, the tumor, would be expected to reduce the body surface voltage generated by the radially activated right ventricular free wall, again unmasking left ventricular forces. This is in accord with the Brody effect (8), which defines the relation between conductivity of intracavitary media, direction of cardiac activation and electrocardiographic potentials.

Echocardiographic findings. While in the past cardiac tumors were diagnosed mostly at autopsy, the availability of echocardiography has enabled the detection of most intracardiac tumors during life. Although diagnosis of intraventricular tumors in infants using M-mode echocardiography has been reported (9–11), precise definition of the tumors with this technique was not possible. Real-time two-dimensional echocardiography permits a more accurate localization, and it provides more information about the characteristics of the tumors (4,12). In our case, scanning in the four chamber apical view demonstrated a massive cloud of echoes that filled almost the entire right ventricular cavity and displaced the ventricular septum against the free wall of the left ventricle. The tricuspid valve was also involved by the tumor mass exhibiting decreased motion and backward displacement toward a small right atrium. Of further interest was that the pulmonary blood flow was mainly dependent on the patency of the ductus arteriosus. Therefore, infusion with prostaglandin E_1 was initiated while the operating room was prepared. At surgery, even though the tumor was not fully resectable because of its location, an attempt was made to facilitate right ventricular function by removing the portion of the tumor protruding into the right ventricular cavity. The small amount of residual right ventricular muscle was not able to function adequately as a pump.

Figure 4. A, Light microscopic section of the tumor showing elongated and stellate cells in a myxoid background. **B**, Electron microscopic section showing mesenchymal cells surrounded by amorphous matrix and collagen fibers.





Light and electron microscopic examination of the tumor. This demonstrated features consistent with a diagnosis of fibroma (13–18). The immaturity of the tissue caused some difficulty in diagnosis, but use of 14 special stains showed the features of an immature fibroma. The tumor was composed mainly of relatively immature mesenchymal tissue without identifying features of cellular differentiation. Although some areas of the tumor were indistinguishable from a myxoma, the intramural location of the tumor (13) and the lack of factor VIII-related antigen staining of the great majority of the cells (19) were not supportive of the diagnosis of myxoma.

Implications. Although intracardiac tumors are very rare in children, they should be included in the differential diagnosis of cyanotic cardiac anomalies. Echocardiography, particularly two-dimensional echocardiography, is an invaluable tool in the prompt diagnosis of intracardiac tumors. As further experience is gained with the technique, real-time images may contain enough information in selected cases to allow for surgical excision of the tumor without further invasive procedures.

We thank Vera E. Paschal for manuscript preparation.

References

1. Griffiths GC. A review of primary tumors of the heart. Prog Cardiovasc Dis 1965;7:465–79.
2. Leach WB. Primary neoplasm of the heart. Arch Pathol 1947;44:198–204.
3. Ilbawi M, DeLeon S, Riggs T, Hernandez R. Primary vascular tumor of the heart in infancy. Chest 1982;81:511–2.
4. Spooner EW, Farina MA, Shaher RM, Foster ED. Left ventricular rhabdomyoma causing subaortic stenosis—the two-dimensional echocardiographic appearance. Pediatr Cardiol 1982;2:67–71.
5. Schmaltz AA, Apitz J. Primary rhabdomyosarcoma of the heart. Pediatr Cardiol 1982;2:73–5.
6. Aryanpur I, Nasarian I. Calcified right ventricular fibroma causing outflow obstruction: report of a case with successful excision. Am J Dis Child 1976;130:1265–7.
7. Chandraratna PAN, San Pedro S, Elkins RC, Grantham N. Echocardiographic, angiocardiographic and surgical correlations in right ventricular myxoma simulating valvar pulmonic stenosis. Circulation 1977;55:619–22.
8. Brody DA. A theoretical analysis of intracavitary blood mass influence on the heart-lead relationship. Circ Res 1956;4:731–8.
9. Allen HD, Blieden LC, Stone FM, Bessinger FB, Lucas RV. Echographic demonstration of a right ventricular tumor in a neonate. J Pediatr 1974;84:854–6.
10. Farooki ZQ, Henry JG, Arciniegas E, Green EW. Ultrasonic pattern of ventricular rhabdomyoma in two infants. Am J Cardiol 1974;34:842–4.
11. Milner S, Abramowitz JA, Levin SE. Rhabdomyoma of the heart in a newborn infant. Diagnosis by echocardiography. Br Heart J 1980;44:224–7.
12. Biancaniello TM, Meyer RA, Gaum WE, Kaplan S. Primary benign intramural ventricular tumors in children. Pre- and postoperative electrocardiographic, echocardiographic and angiocardiographic evaluation. Am Heart J 1982;103:852–7.
13. Geha AS, Weidman WH, Soule EH, McGoon DC. Intramural ventricular cardiac fibroma. Circulation 1967;36:427–40.
14. Folger GM, Peters HJ. Nodular fibroelastosis (fibroelastic hamartoma): a tumorous malformation of the heart. Am J Cardiol 1968;21:420–7.
15. Harpin NJ, Wilson JM, Grau FG, Gay WA. Experience with primary tumors of the heart: clinical and pathological study of seventeen cases. Johns Hopkins Med J 1974;134:141–55.
16. Feldman PS, Meyer MW. Fibroelastic hamartoma (fibroma) of the heart. Cancer 1976;38:314–23.
17. Gonzalez-Crussi F, Eberts TJ, Mir in D. Congenital fibrous hamartoma of the heart. Arch Pathol 1978;102:491–3.
18. Fine G, Osamura RY, Lee MW. Ultrastructure of the myocardial fibroma. Arch Pathol 1979;103:11–17.
19. Morales AR, Fine G, Castro A, Nadji M. Cardiac myxoma (endocardioma): an immunocytochemical assessment of histogenesis. Hum Pathol 1981;12:896–9.